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Saleem Abdulkarim

*Aga Khan University*, [saleem.abdulkarim@aku.edu](mailto:saleem.abdulkarim@aku.edu)

Fredrick Otieno

*Aga Khan University*, [fredrick.otieno@aku.edu](mailto:fredrick.otieno@aku.edu)

Sayed Karar

*Aga Khan University*, [sayed.karar@aku.edu](mailto:sayed.karar@aku.edu)

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# Adult-onset Still's disease triggered by pregnancy

Saleem Abdulkarim, MBChB<sup>a</sup>, Fredrick Otieno, MBChB<sup>b</sup>, and Sayed K. Ali, MD, FACP<sup>a</sup> 

<sup>a</sup>Section of Internal Medicine, Department of Medicine, Aga Khan University, Nairobi, Kenya; <sup>b</sup>Section of Rheumatology, Department of Medicine, Aga Khan University, Nairobi, Kenya

## ABSTRACT

Adult-onset Still's disease (AOSD) is an inflammatory disorder with arthritis, quotidian fever, evanescent rash, elevated white cell count, elevated ferritin, and abnormal liver function tests. The diagnosis requires the exclusion of other inflammatory, infectious, and malignant conditions. Onset of AOSD, though rare, has been known to occur in pregnancy. We present a 28-year-old woman diagnosed with AOSD at 3 months gestation.

**KEYWORDS** Pregnancy; Still's disease; treatment

Adult-onset Still's disease (AOSD) is an inflammatory condition characterized clinically by quotidian fever, arthritis, and an evanescent rash associated with an elevated white cell count with neutrophilia, elevated ferritin levels, and abnormal liver function tests. It is a rare condition with an incidence reported in a retrospective French study of 0.16 per 100,000.<sup>1</sup> The incidence of onset of AOSD in pregnancy is even less frequent, with a literature review revealing 20 reported cases to date.<sup>2</sup> The exact etiopathogenesis of the condition is not clear, but a variety of infectious causes on a background of genetically predisposed individuals is believed to play a role.<sup>3–7</sup> The diagnosis of AOSD requires exclusion of inflammatory, infectious, and malignant conditions. The Yamaguchi criteria, which include clinical and laboratory parameters, are commonly used to help make the diagnosis.<sup>8</sup>

## CASE PRESENTATION

A 28-year-old woman, para 2 + 0 gravida 3, at 12 weeks gestation by dates with no prior medical comorbidities presented to our institution with a 2-week history of joint pain. She noted pain, swelling, and stiffness of the ankles, knees, wrists, elbows, and shoulders bilaterally with associated limitation of movement. She also reported daily fever with onset in the evening and a sore throat. She denied having a rash, oral ulcers, or alopecia. She had completed two courses of oral antibiotics (amoxicillin/clavulanic acid and cefuroxime) for presumed infection at an external facility where she was

noted to have fevers of up to 39°C with no clear focus of infection. On examination, she had tender joints in the distribution described but no palpable hepatomegaly or splenomegaly. Her hemoglobin was 11.2 g/dL; white cell count,  $19.13 \times 10^9/L$  (neutrophils 85%); platelets,  $322 \times 10^9/L$ ; C-reactive protein, 363 mg/L; erythrocyte sedimentation rate, 60 mm/h; procalcitonin, 0.25 ng/mL; ferritin, 5306 ng/mL; and antinuclear antibodies, rheumatoid factor, and anticyclic citrullinated peptide were negative. A complete metabolic panel, including liver function tests, was normal. Blood and urine cultures were negative. Tests for malaria, syphilis, dengue virus, leptospirosis, rickettsia, West Nile virus, and Chikungunya virus were also negative. An HIV test was nonreactive. The patient was subsequently started on steroids with intravenous methylprednisolone at 250 mg with improvement of her symptoms. She did not receive intravenous immunoglobulin. She was discharged on day 2 with complete resolution of her fevers and joint pain on prednisone 20 mg twice a day.

She returned to our institution 9 weeks later (at 21 weeks gestation) and was noted to be jaundiced and have abnormal liver function tests (alanine aminotransferase, 1137 U/L; aspartate aminotransferase, 974 U/L; alkaline phosphatase, 173 U/L; gamma glutamyl transferase, 564 U/L; total bilirubin, 210  $\mu\text{mol/L}$ ; direct bilirubin, 200  $\mu\text{mol/L}$ ). Hepatitis A, B, and C serologies were negative. An autoimmune hepatitis panel, including anti-smooth muscle, anti-mitochondrial, and anti-ds-DNA antibodies, was also negative. Magnetic retrograde cholangiopancreatography was normal. This was

**Corresponding author:** Sayed K. Ali, MD, FACP, Section of Internal Medicine, Department of Medicine, Aga Khan University, P.O. Box 30270-00100, Nairobi, Kenya (e-mail: [Sayed.karar@aku.edu](mailto:Sayed.karar@aku.edu))

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considered a flare-up of her AOSD. She was started on intravenous methylprednisolone 500 mg once a day with improvement in her liver function tests (alanine amino transferase, 897 U/L; aspartate aminotransferase, 878 U/L; alkaline phosphatase, 143 U/L; gamma glutamyl transferase, 476 U/L; total bilirubin, 183  $\mu$ mol/L; direct bilirubin, 175  $\mu$ mol/L). She was discharged on day 3 with prednisone 20 mg once a day and azathioprine 50 mg once a day. She was doing well during follow-up at 38 weeks gestation with normal liver function. Her maintenance medications included prednisone 10 mg twice a day and azathioprine 100 mg once a day.

## DISCUSSION

AOSD onset during pregnancy remains rare.<sup>2</sup> AOSD demonstrates a bimodal age distribution with peaks at 15 to 25 and 36 to 46 years of age.<sup>9</sup> The age distribution in a case series published locally was between 21 and 48 years (mean age, 35).<sup>9</sup> The pattern of presentation of our patient was similar to that reported in a case series of patients with onset of AOSD in pregnancy with fever, arthralgia, and sore throat<sup>2</sup>; 80% of those patients were noted to have a rash, which was not noted in our patient, though the rash may be more difficult to detect in dark-skinned individuals. Of note is that all patients in the series had systemic involvement, and none presented with a chronic articular form. The gestational age of onset was noted to be in the first or second trimester (8 to 26 weeks), with our patient having onset of symptoms at 10 weeks with a diagnosis made at 12 weeks. Ferritin levels were elevated in all patients, ranging from 1311 to 41,424 ng/mL. Flares in the patients appeared between the first and second trimesters, earlier than in patients with preexisting AOSD.<sup>10</sup> Obstetric complications seemed to be common, with 50% of the women having preterm births and 15% complicated by intrauterine growth restriction.<sup>10</sup>

Treatment of AOSD can be challenging. Steroids are usually utilized but carry the risk of gestational diabetes, hypertension, and frequent monitoring. Intravenous immunoglobulin may be used in life-threatening cases.<sup>11</sup> Azathioprine is sometimes preferred due to its steroid-sparing

property.<sup>12</sup> Cases reported of the condition in pregnancy remain few and treatment experience so far is derived from case series.

## ORCID

Sayed K. Ali  <http://orcid.org/0000-0003-0750-2903>

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